DRÜSEN OF THE OPTIC DISK – HEMORRHAGIC COMPLICATIONS*

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DRÜSEN (HYALINE BODIES, CONCRETIONS) of the optic disk have been well recognized both clinically and histologically for over 100 years. For many years their clinical importance was the ophthalmoscopic appearance in the differential diagnosis of papilledema.

Lorentzen[1] in 1966 wrote a comprehensive monograph on optic-nerve drüsen, emphasizing the clinical and the genetic aspects. For an extensive and complete bibliography of over 200 references of optic-nerve drüsen, the reader is referred to his study. For an excellent discussion of the clinical aspects, particularly as to diagnosis, the reader is referred to the textbook of Walsh and Hoyt.[2]

For many years it was thought that these lesions were completely innocuous. Although the field defects arising from this condition were first described some fifty years ago, only relatively recently have they been recognized as a characteristic clinical finding.

Lauber[3] in 1921, was among the first to describe the field defect. He found visual-field defects in 9 out of 14 examined cases (64 per cent). It was not until Rucker's[4] report in 1944 that this defect was again emphasized. Since that time there have been a number of reports on the field defects in this condition.[5,6,7] In 1957, Lansche and Rucker[8] reported a visual-field study of 14 patients with repeated examinations over a number of years. Of the 14 patients, only 6 had a slowly progressive field defect. To date, the largest and the most complete study of the visual fields in this condition is that of Lorentzen[1] who reported a total of 91 visual fields with 40 subjects having bilateral defects and 11 subjects having unilateral defects.

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This work was supported in part by USPHS Research Grant 1–R01–NB08033 from the National Institutes of Neurological Diseases and Stroke, Bethesda, Maryland.

TR. AM. OPHTH. SOC., vol. 68, 1970

He felt there were pathologic fields present in 79 (87 per cent) of these patients. However, it has been stated[8,9] that very rarely does a patient lose central vision from optic-nerve drüsen.

Hemorrhage of the optic disk secondary to giant drüsen is a rare condition. We have been able to find only nine recorded cases in the literature. With one exception, these cases are scattered throughout reports on the condition, and are mentioned only as incidental findings. The first and apparently only specific report on hemorrhage associated with drüsen of the disk did not appear until 1967, when Gaynes and Towle[10] reported a case of a 19-year-old female with vitreous hemorrhage developing from a disk with drüsen during a migraine attack.

The first recorded case of vitreous hemorrhage in a case report of giant drüsen was mentioned by Gifford[11] in 1895. The next example was not reported until 1940 when Reese[12] described a young patient with drüsen of the optic disk followed for a number of years prior to a sudden blurring of vision from vitreous hemorrhage over the disk. Lorentzen[1] states that no instance of hemorrhage occurred in his series of 70 patients but he does mention three instances from the French literature. In Gallais's patient[13] a minor peripapillary hemorrhage of the left disk was noted in an 11-year-old girl. The hemorrhage had disappeared on re-examination three weeks later. Lorentzen also states that Brégeat[14] noticed a streaky hemorrhage on the disk in two patients with drüsen. In both of these, the hemorrhage had disappeared on subsequent examination. Lorentzen[1] also described a young man he saw in Professor Paufique's department in Lyon in 1959 with a vitreous hemorrhage over a disk with drüsen. In Walsh and Hoyt's textbook,[2] two cases are mentioned in a footnote. In these, splinter hemorrhages were seen on the disk margin in patients with drüsen. They state that the importance of this finding is the increased difficulty in the differential diagnosis of papilledema.

Lorentzen[1] summarizes the hemorrhage associated with drüsen as follows: "Minor hemorrhages on or close to the optic disc and even in the vitreous body, may thus occur in association with drüsen of the optic disk. Such are, in the author's opinion, so rare that instead of regarding them primarily due to the drüsen, one should pay special attention to them when present." In other words, another possible etiology should be suspected.

From the above survey of the literature it is apparent that hemorrhage in and around drüsen of the optic disk is not only a rare

condition, but is one that has been almost completely ignored in the ophthalmic literature. We believe, therefore, that the present series of seven cases is worth recording, not only to give a description of the condition, but also to discuss some possible relations of hemorrhage to the well-recognized field defects of this condition. In addition, we would like to emphasize occurrence of deep peripapillary hemorrhage, which apparently has not been previously described. Also included are the first histopathologic descriptions of hemorrhage in relation to drüsen of the optic disk.

REPORT OF CASES

The following series of seven cases of hemorrhage associated with drüsen of the optic disk has been collected by the authors from various sources. Cases I and II are from the private practice files of one of the authors of this paper (TES). Case III was presented by Dr Frank Winter at the meeting of the Verhoeff Society in 1967. Case IV is from the collection of autopsy material of the Ophthalmic Pathology Laboratory of Washington University. Case V is contributed by Dr Allan E. Kolker of the Department of Ophthalmology, Washington University. Case VI was a patient seen by one of us (AJG) in the Eye Consult Office, Department of Ophthalmology, Washington University School of Medicine. Case VII was a patient seen and diagnosed by one of the authors (MN) in the Eye Clinic of the Jewish Hospital of St. Louis.

CASE I

A 58-year-old white female was admitted to Barnes Hospital because of a blind, painful left eye from pupillary block glaucoma secondary to an old anterior uveitus. An iridectomy was done with uneventful postoperative course. Some two years later she had a recurrence of the inflammation with pain, and the eye was enucleated.

On first examination, vision in the right eye was 20/20 uncorrected. The anterior segment was normal. On ophthalmoscopic examination the fundus was normal except for the disk which was elevated about one diopter with slightly blurred margins. The fields showed an almost complete inferior loss, tending to be arcuate (Figure 1A). Intraocular tension was 16 mm Hg.

Because of the field loss and the appearance of the disk, the presence of giant drüsen of the disk was suspected even though actual concretions were not noted. The patient stated that about eight months prior to admission she had been under the care of another ophthalmologist who had advised medical and neurological consultation. On reviewing his records, it was found that the right disk was blurred and elevated with some hemorrhage on its surface. There was marked nasal field loss at this time.

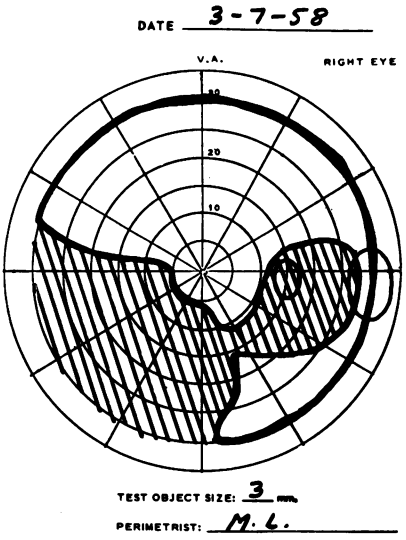

FIGURE 1A

(Case 1) Inferior arcuate defect in right eye when first examined.

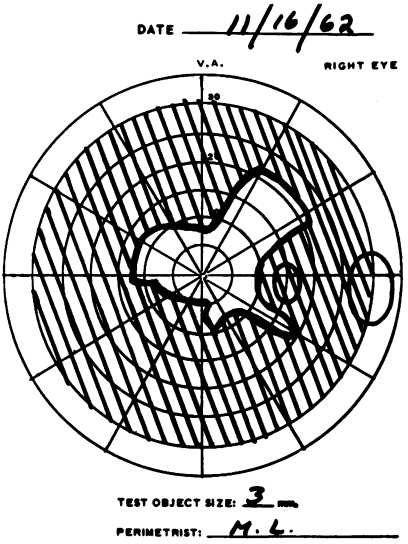

FIGURE 1B

(Case 1) Superior field loss in right eye associated with subretinal peripapillary hemorrhage.

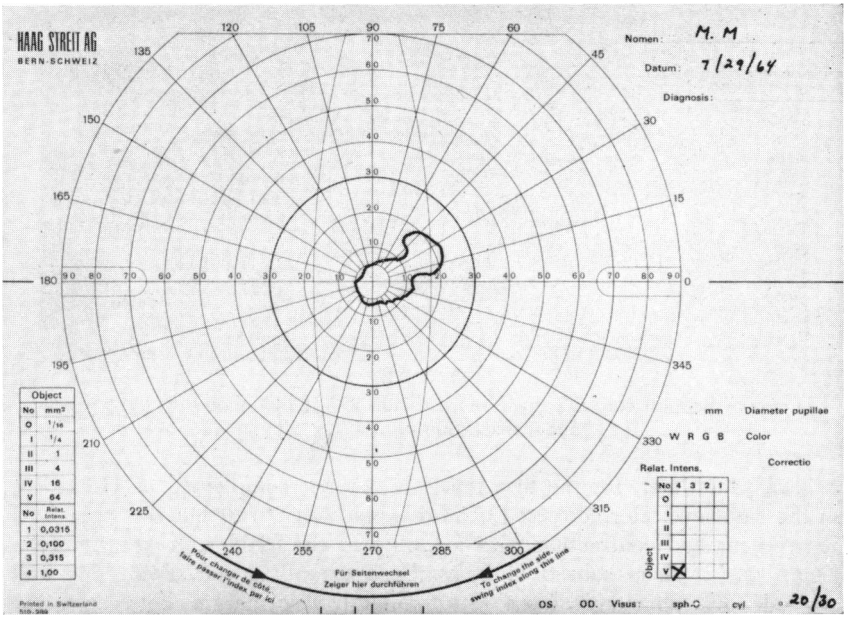

FIGURE 1C

(Case 1) Advanced contraction of the field in the right eye seven years following initial examination.

The patient was followed regularly for the next two and one-half years. Fields showed very little progression but there was an arcuate defect suggestive of glaucoma. Repeated tonograms were within normal limits. The disk remained about the same in appearance except that concretions were noted definitely for the first time.

About this time, the histologic examination of the enucleated left eye confirmed the presence of many concretions in the disk on the opposite side (Figure 2). This examination revealed the disk to be very cellular, probably because of proliferation of glial cells. There was a thin fibrovascular membrane over the disk surface and some retinal edema around the disk. The choroid adjacent to the disk showed some non-specific inflammation. These findings were unusual and quite different from those seen with uncomplicated drüsen. They were consistent with hemorrhage, or possibly associated with the chronic anterior uveitis.

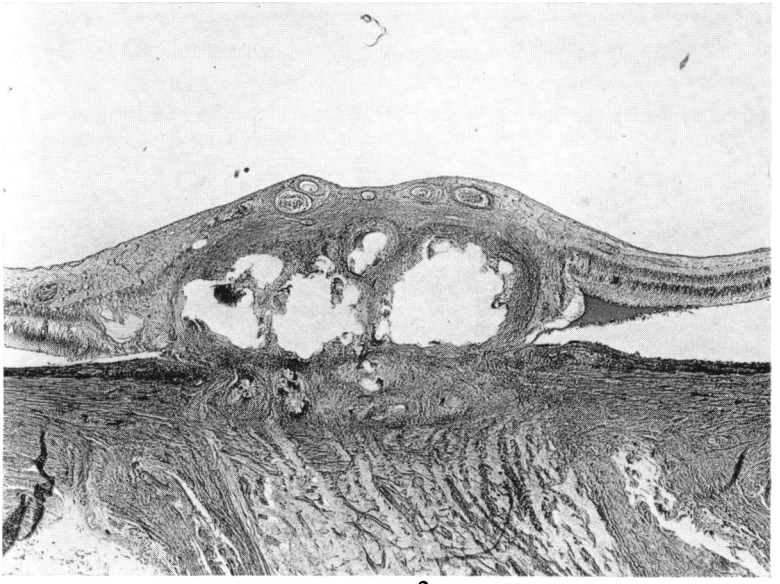

FIGURE 2

(Case 1) Section through optic nerve head of left eye showing giant drüsen and surrounding gliosis. (× 28)

Two years later, for the first time, the patient complained of visual loss in the right eye. Her corrected central vision was 20/20 but the right field showed marked contraction with a baring of the blind spot (Figure 1B). There was a deep subretinal hemorrhage extending out about one and one-half disk diameters from the temporal disk margin between eight o'clock and ten o'clock. There was some attenuation of the vessels and arteriovenous compression.

Six months later, her central vision had decreased to 20/25 and she was still complaining of continued visual loss. Her fields showed a marked contraction inside eight degrees (Figure 1C), but the hemorrhage around the disk had completely disappeared.

Because of the field loss, the patient was admitted to the hospital for a complete examination. Ophthalmological findings were normal except for extreme field defect and the appearance of the disk. Neurological examination also was normal.

About six months later, her central vision had decreased to 20/40 and her field was concentrically contracted to about five degrees. She was again admitted to the hospital on the Neurosurgical Service for angiograms, which were normal. At this time, the concretions of the disk were very obvious, and the disk appeared to be more pale.

When the patient was last seen two years later, her field was extremely contracted, central visual acuity was 20/40+ and there were still obvious concretions of the disk. The patient had a severe cardiac attack and has not been able to return for examination. She reports that she cannot read, watches television with difficulty, and her local physician states that her central vision is 20/100 with a field of about five degrees.

Comment

This patient with marked progressive field loss in her only eye probably represents one of the few cases of extreme visual loss resulting from drüsen of the disk. Diagnosis was made on clinical evidence substantiated by the histological report of the presence of concretions in the opposite eye. Extensive examination revealed no other reason for the visual loss.

Apparently there were two episodes of hemorrhage. The hemorrhage superficial to the disk was noted in the records of another ophthalmologist some eight months prior to our first examination. Approximately four and one-half years later the second hemorrhage was noted extending subretinally around the disk. Coincidentally, there was a rapid loss of visual field, which progressed to a very small residual field with marked loss of central vision.

CASE II

A 9-year-old white male was first seen for an opinion as to the condition of the optic disks. He had no visual complaints. A neurosurgeon had made a diagnosis of papilledema and advised an air study. Two other neurosurgical consultants felt that the appearance of the disks was not evidence enough for this.

Our examination revealed the patient's vision to be 20/20. His eyes were completely normal except for the disks, which were elevated about two diopters with steep margins. The diagnosis of drüsen was made, and the

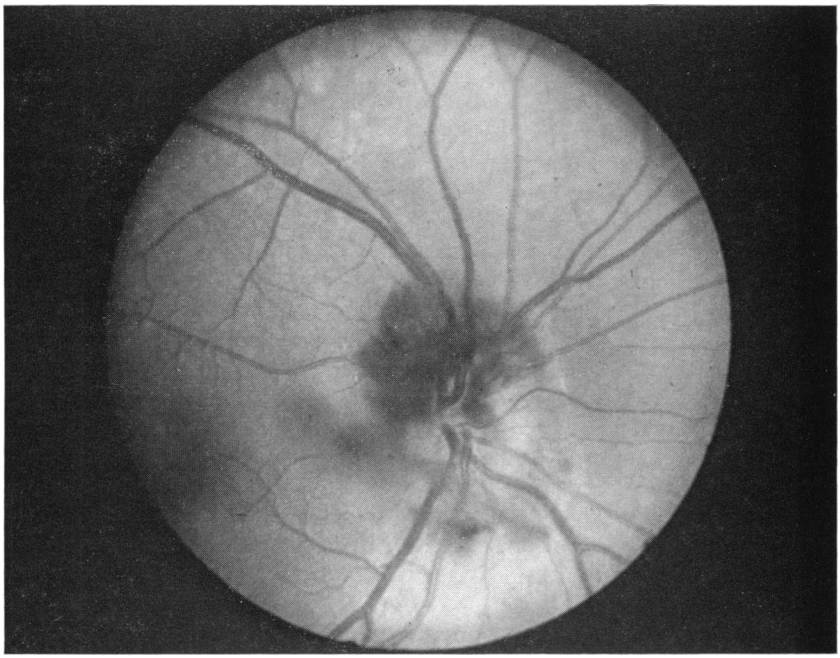

FIGURE 3A

(Case II) Right optic nerve head with overlying superficial hemorrhage that had ruptured into the vitreous.

patient was followed for two years with no change in his condition except that drüsen became more prominent.

At age 16, the patient was seen again for sudden blurring of vision in the right eye, which had occurred the previous evening following a game of tennis. He described a gray area to the right of fixation, which had become larger, developing into a black spot that obscured his vision. Examination showed a large fresh hemorrhage on the disk, particularly in the upper half (Figure 3A). Much of this was preretinal, with some strands extending into the vitreous. The left disk was unchanged (Figure 3B).

Visual fields were not studied, but a complete medical and hematological examination was negative. In three weeks, the majority of the hemorrhage had disappeared except for a superficial fleck at nine o'clock and a rim of deep hemorrhage around the disk margin superiorly (Figure 3C).

One week later, the patient had a recurrence of blurred vision, but examination showed his vision to be still 20/20. There was a fresh hemorrhage in the upper quadrant of the same disk apparently at the same spot as the first one. This tended to spread out along the fiber layers of the retina and had a long thin plume extending into the vitreous.

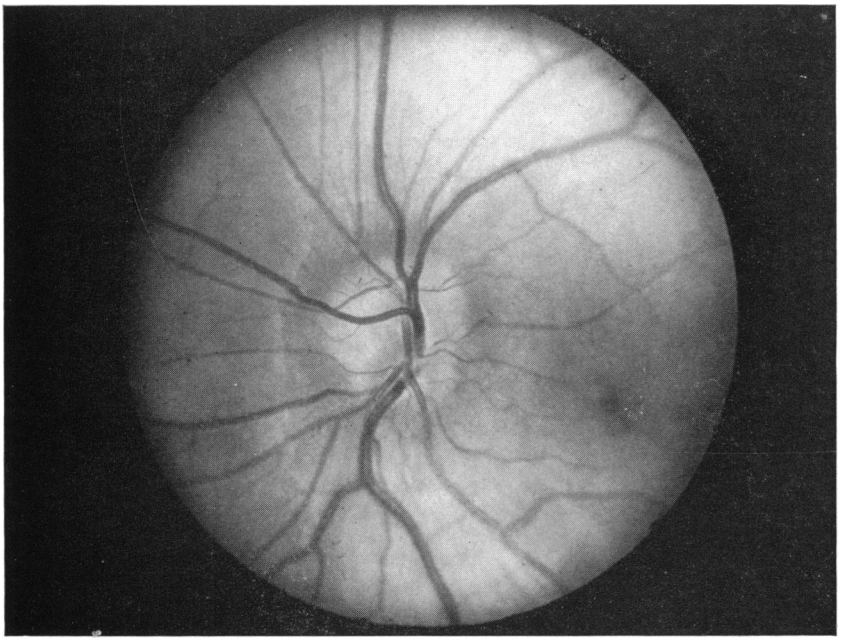

FIGURE 3B

(Case II) Left eye showing blurred disk margin and nasal crescentic reflex secondary to drüsen.

One month later, the patient had a slight recurrence of blurring but has not been re-examined. He has had no further symptoms in the right eye and has been inducted into the Armed Services.

Comment

A vitreous hemorrhage developed in this patient, secondary to drüsen of the disk. This vitreous hemorrhage occurred a number of years after the original diagnosis of drüsen of the disk was made. This situation is similar to that of the patient reported by Reese.[12] It is interesting to note that his original episode came on immediately after strenuous exertion during a game of tennis. It is possible that this could be an exacerbating factor. It was thought that an attack of migraine was a similar factor in the case reported by Gaynes and Towle.[10] This patient demonstrates that with age the drüsen in the disk becomes more easily seen ophthalmoscopically.

CASE III

This patient was presented by Dr Frank Winter at the 1967 meeting of

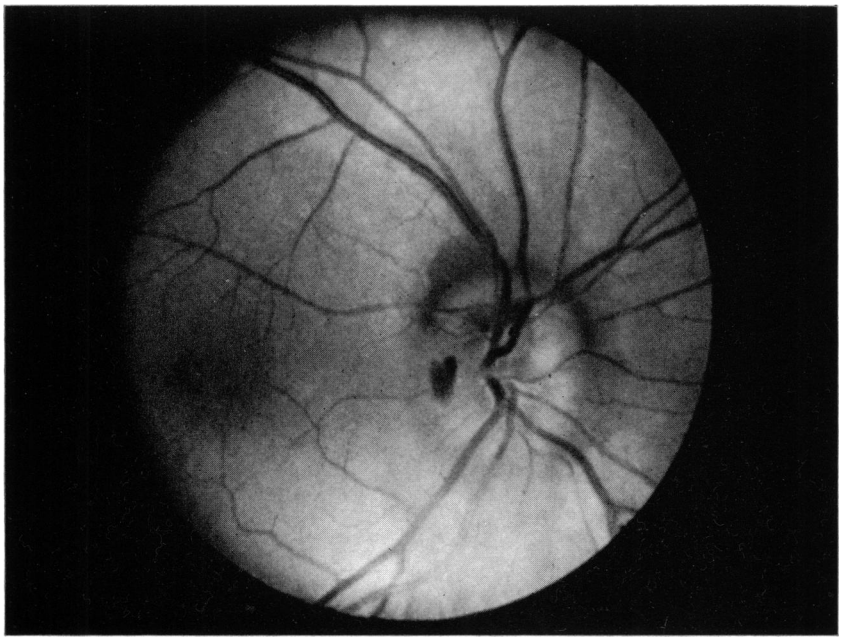

FIGURE 3C

(Case II) Right eye three weeks later: hemorrhage has partially resorbed. Note small splinter hemorrhage disk surface inferotemporally.

the Verhoeff Society and is included with his permission. The contributing surgeon has been dead for some time and his office files are not available. Therefore, the only history available is that in the Ophthalmic Pathology Laboratory of Stanford University.

A 12-year-old male consulted an ophthalmologist because of blurred vision in the left eye of several weeks' duration. On examination, a discrete elevated pigmented mass was seen deep under the retina between the disk and the fovea. A diagnosis of malignant melanoma was made by several observers and the eye was enucleated.

The histologic specimen showed a disk filled with deep-staining drüsen of varying size (Figure 4A). Between the macula and the disk there was a discrete subretinal pigmented mass lying between the outer retinal layers and the pigment epithelium (Figure 4B). This was made up of old hemorrhage. No hemorrhage was seen in the disk, but at the temporal margin of the disk there was a strand of fibrous tissue with old hemorrhage extending from the disk under the retina toward the mass (Figure 4C).

Comment

This patient had a deep subretinal hemorrhage forming a hematoma

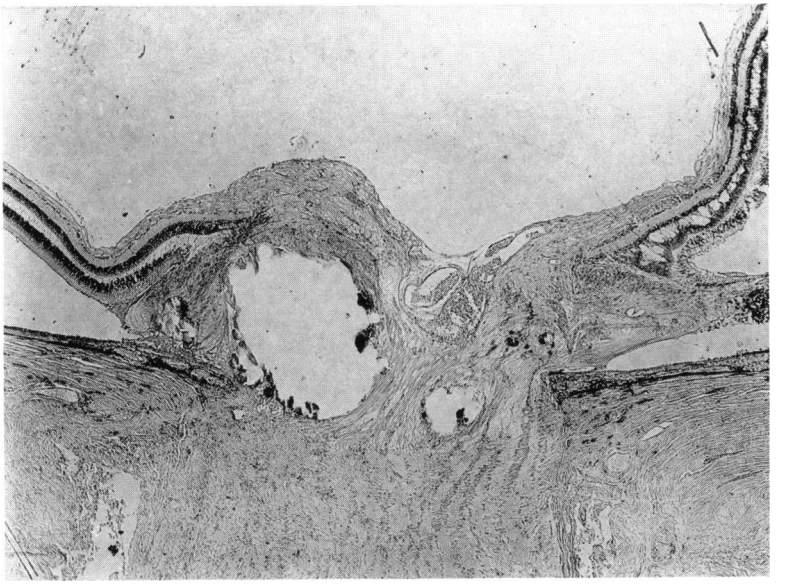

FIGURE 4A

(Case III) Section through disk showing large drüsen nasally, smaller drüsen temporally. (× 28)

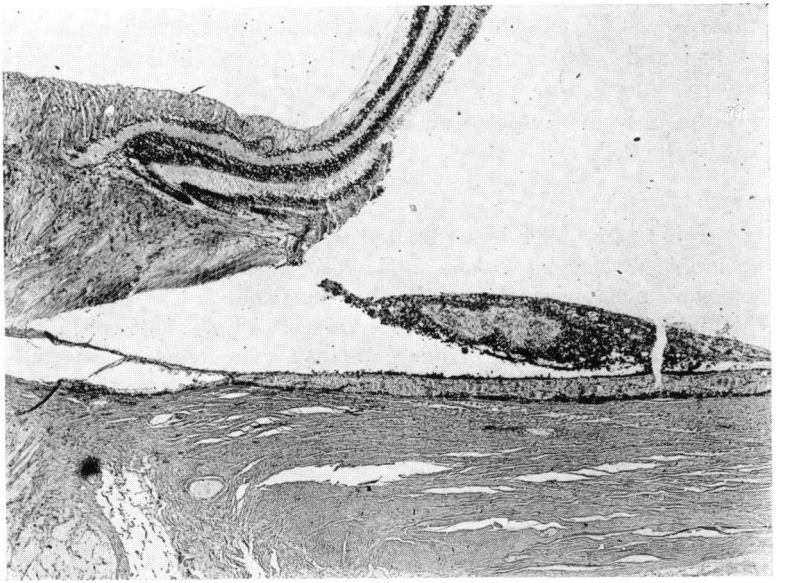

FIGURE 4B

(Case III) Section showing subretinal macular hematoma. (× 33)

FIGURE 4C
(Case III) Section showing strand of fibrous tissue with old blood extending subretinally from temporal margin of disk.

simulating a malignant melanoma. Enucleation occurred a number of years ago and with modern techniques, such as fluorescein angiography, the enucleation might have been avoided. In our experience, this is the only eye enucleated because of hemorrhage secondary to drüsen of the disk.

CASE IV

A 62-year-old white male was admitted to the hospital for the fourth time with chronic myelocytic leukemia. He died several days after admission and a complete autopsy including the eyes was done.

The first admission was about three years previously, following a diffuse hemorrhage after extraction of teeth. He had been found to have splenomegaly with a white blood count of 575,000. The diagnosis of chronic myelocytic leukemia was made after examination of the bone marrow. The patient had had various treatments and had done well until his final admission.

About a year and one-half after onset the patient was admitted to the hospital for the second time because of a sudden loss of vision in his left eye found to be caused by a vitreous hemorrhage. At the last admission it was said that his vision had never completely improved. At this time,

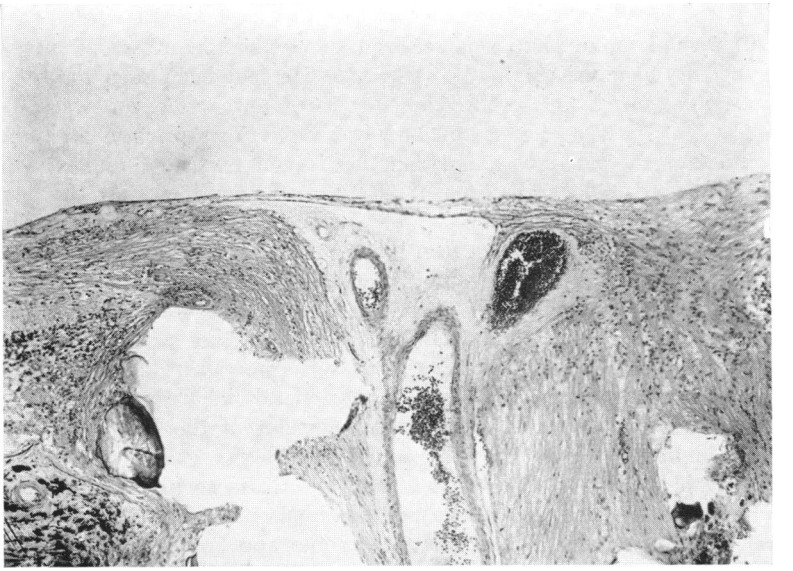

FIGURE 5
(Case IV) Section through disk. Fibrovascular membrane overlying physiologic cup. (× 63)

floaters were noted in the left eye, the disk was blurred, and there was some "patchy exudate nasal to disk."

Histologic examination showed many large drüsen in the disk filling almost all of the nerve head; the nerve fibers were normal. On the surface of the disk there was a thin fibrovascular proliferation that formed a membrane on the surface of the disk over the physiologic cup (Figure 5).

Comment

Clinically, this patient resembles our other patient (Case II) as well as several other patients previously reported. With the chronic myelocytic leukemia, he may very well have had a hemorrhagic diathesis. However, the clinical appearance of the hemorrhage resembled that of drüsen rather than that usually seen in leukemia. Apparently this patient did not completely recover his vision following the hemorrhage. This patient had, we believe, the only instance of vitreous hemorrhage from drüsen that has been examined histologically. The thin fibrovascular membrane on the surface of the disk probably represents organized remnants of the hemorrhage.

CASE V

A 16-year-old white male was referred for evaluation of visual loss and peripapillary hemorrhage in the right eye. He had been seen for routine eye examinations by his ophthalmologist for five years. Vision was 20/20 in each eye. His last eye examination was three years prior to his present episode. For several days the patient had noted persistent blurred vision and was found to have a vision of 20/30 in the right eye. At the lateral margin of the right disk was an elevated purplish-colored mass. Three days later his vision had decreased to 20/70 and it was noted that around this elevation was a large area of hemorrhage with folds extending into the macula. The possibility of a malignant melanoma was considered and the patient was referred to Dr Allan E. Kolker for further evaluation.

Examination showed visual acuity without correction to be 20/30 in the right eye and 20/15 in the left eye. Peripheral fields were normal, but central fields demonstrated bilateral enlargement of the blind spot in an arcuate pattern (Figure 6). The visual-field defect extended to fixation in the right eye but not in the left eye. Ophthalmoscopic examination of the right eye revealed the disk to be of normal color with blurred margins superiorly and nasally. There was a refractile appearance such as is seen with drüsen at the ten o'clock and three o'clock positions on the disk. Surrounding the disk superiorly was a zone of deep, dark-colored hemorrhage, which extended temporally toward the macula with radiating folds of edema above it (Figure 7A). Ophthalmoscopic examination of the left eye showed a disk of normal color with an area of elevation and drüsen at ten o'clock and at three o'clock. Surrounding the disk superiorly and nasally from about nine o'clock to two o'clock was an area of chorioretinal atrophy (Figure 7B). On several follow-up examinations the vision gradually improved and the hemorrhage was absorbed.

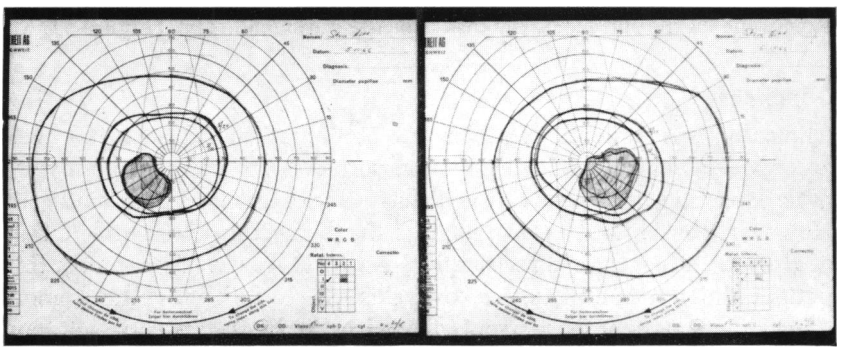

FIGURE 6

(Case v) Visual fields, right eye: relative enlargement of blind spot extending in an inferior arcuate pattern toward fixation. Left eye: enlargement of blind spot extending in inferior arcuate pattern. The peripheral isopters are normal.

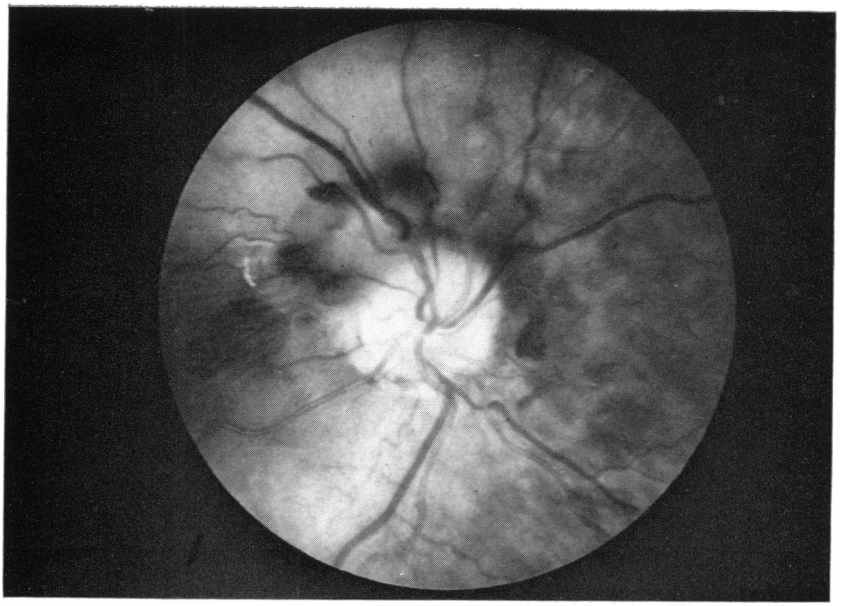

FIGURE 7A

(Case v) A right eye showing deep peripapillary hemorrhage superiorly extending toward the macula.

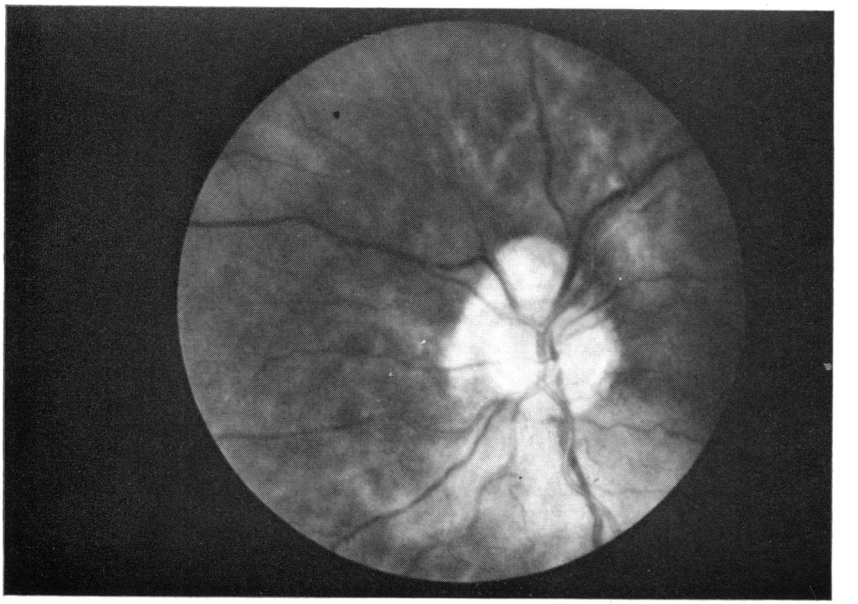

FIGURE 7B

(Case v) Left eye showing area of chorioretinal atrophy superiorly and temporally.

When the patient was examined one year later the vision was 20/20 in both eyes. There were inferior arcuate scotomata in both eyes as in the initial examination. The hemorrhage around the right disk had disappeared and there was beginning peripapillary pigmentary atrophy similar to that seen in the left eye on initial examination.

Comment

Deep peripapillary hemorrhages developed secondary to drüsen with involvement of the submacular area, simulating a malignant melanoma. The uninvolved eye showed a peripapillary atrophy with pigmentation, which possibly resulted from a previous but asymptomatic hemorrhage.

CASE VI

This 26-year-old white male was admitted to Barnes Hospital on the Neurology Service, because of blurred vision in the left eye of two months' duration. He was originally told that he had an "inflammation of his eye" and was treated with steroids.

He was seen by the Eye Consult Service where his visual acuity was found to be 20/25 in the right eye and 20/70 in the left eye. Ophthalmoscopic examination revealed multiple drüsen scattered over each disk surface, more marked in the left eye. Between the disk and the left macula there was a diffuse greenish-gray elevation thought to be a subretinal hemorrhage (Figure 8). There were also two superficial hemorrhages at the superior and inferior margins of the disk. Visual fields of the right eye showed an irregular inferior nasal quadrantal defect and a normal blind spot. In the left eye there was enlargement of the blind spot with some deformity of the inner isopters (Figure 9).

The patient was seen again five years later, at which time vision in the right eye had improved to 20/20 but the vision of the left eye remained unchanged at 20/70. Ophthalmoscopic examination revealed a large chorioretinal scar with diffuse mottled pigmentation in the lower papillomacular bundle up to the fovea. Visual fields revealed a superior arcuate defect extending to fixation.

Comment

This patient represents the third example of macular hemorrhage secondary to drüsen of the optic disk. Five years later there was permanent visual loss from a macular lesion.

CASE VII

This 33-year-old Negro male with a history of distorted vision in the right eye of several months' duration was admitted to the Jewish Hospital of

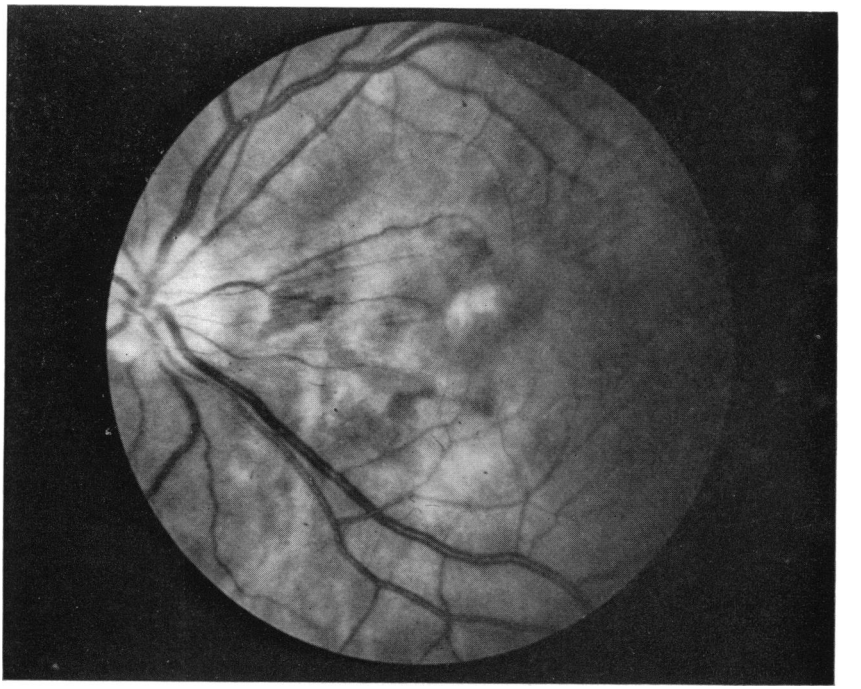

FIGURE 8

(Case VI) Left eye showing drüsen of the disk and extensive subretinal hemorrhage involving the macula.

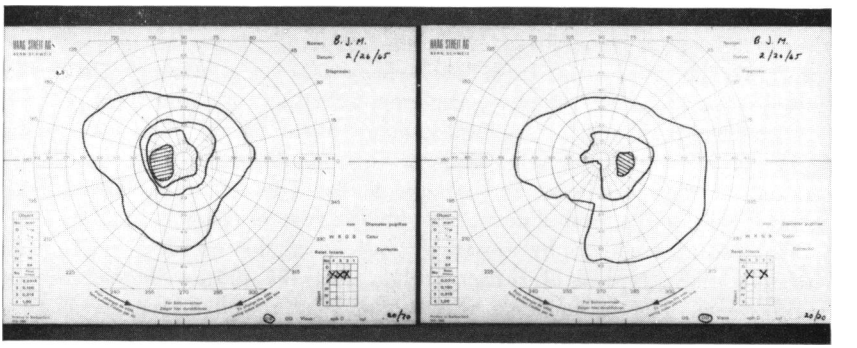

FIGURE 9

(Case VI) Visual fields, right eye: central and peripheral nasal defect and moderate enlargement of blind spot. Left eye: marked enlargement of the blind spot.

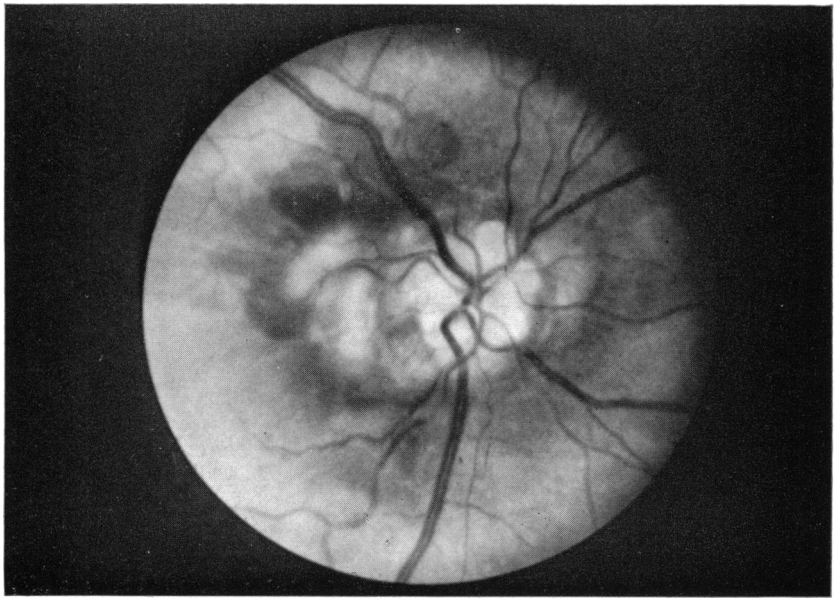

FIGURE 10A

(Case VII) Right eye: giant drüsen of the optic nerve with extensive subretinal hemorrhage temporally.

St. Louis with the diagnosis of papilledema. Medical evaluation, including lumbar puncture and skull roentgenograms, was negative.

The patient was seen in consultation by the eye service. The visual acuity was found to be 20/20 in both eyes and the ophthalmoscopic examination revealed multiple drüsen of the optic nerve heads bilaterally but no papilledema. In the right eye there was an irregular pattern of diffuse subretinal hemorrhage extending temporally from the disk (Figure 10A). In the left eye no fresh hemorrhage was seen, although there was extensive peripapillary atrophy and pigment clumping (Figure 10B).

Visual fields revealed considerable enlargement of the blind spot in the right eye along with an inferior nerve-fiber bundle defect (Roenne Step) present in the peripheral and central fields. The left eye showed both inferior-nasal and temporal nerve-fiber-bundle defects (Figure 11).

Fluorescein angiography revealed clusters of fluorescence in the late arteriole phase typical of drüsen rather than the diffuse fluorescence of papilledema. The hemorrhage was outlined by retrofluorescence (Figure 12).

The hemorrhage gradually resolved and six months later peripapillary atrophy and pigment clumping similar to that seen initially developed in the left eye with no change in vision (Figures 13A and 13B).

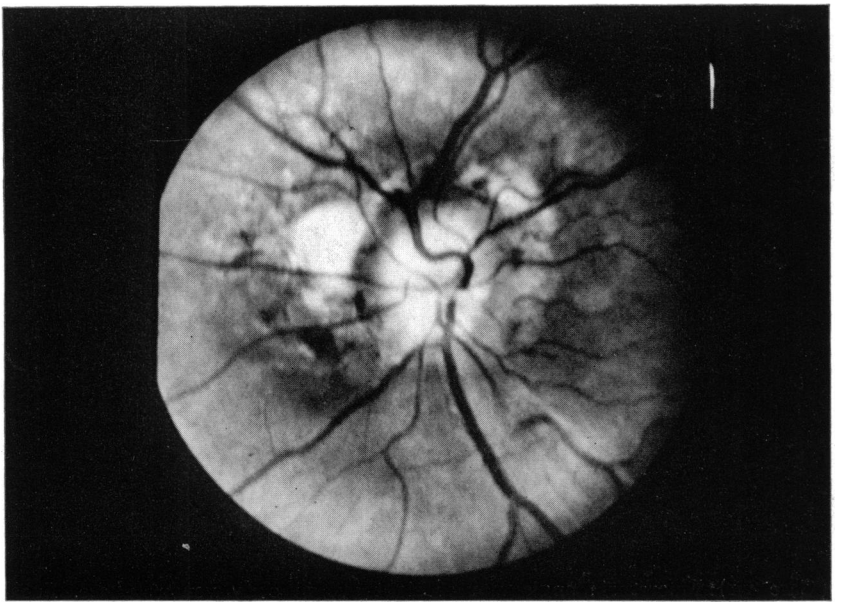

FIGURE 10B

(Case VII) Left eye: areas of extensive peripapillary chorioretinal atrophy and pigment disturbance.

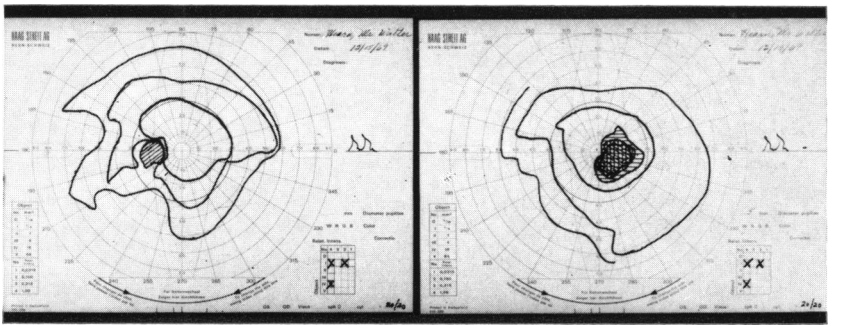

FIGURE 11

(Case VII) Visual fields, right eye: peripheral and central inferior nasal step and relative enlargement of the blind spot. Left eye: a cuneate defect in the inferior temporal field and absolute enlargement of the blind spot.

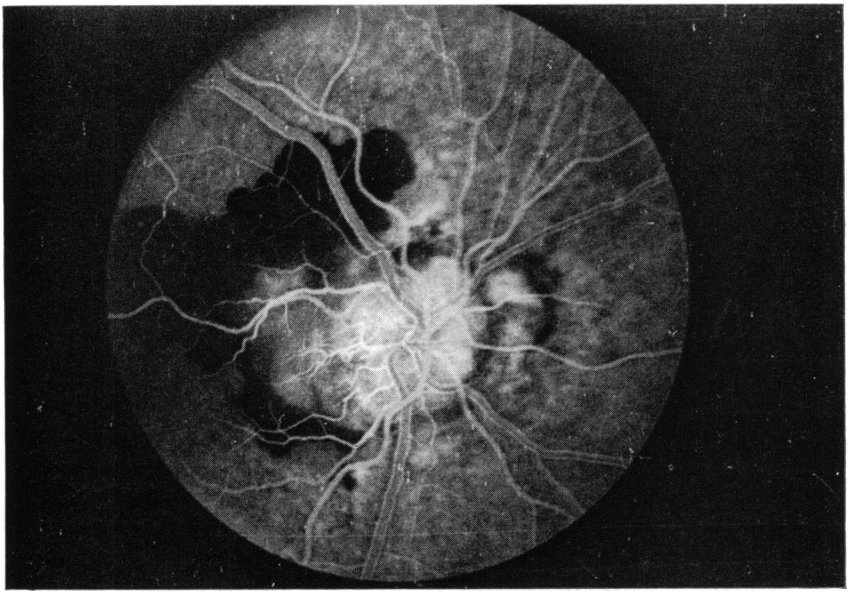

FIGURE 12

(Case VII) Fluorescein angiogram of right eye in early venous phase. Focal fluorescence of the disk because of drüsen with the subretinal hemorrhage outlined by retrofluorescence.

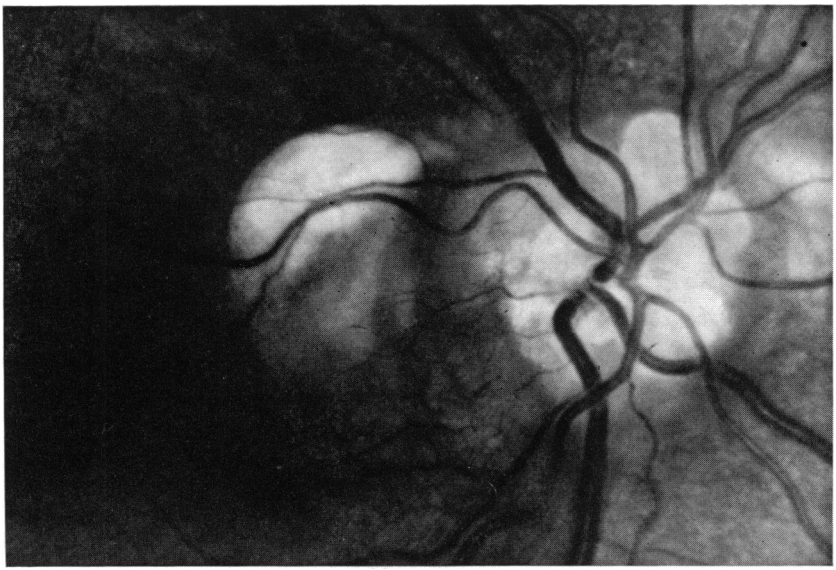

FIGURE 13A

(Case VII) Right eye six months following hemorrhage. Residual peripapillary chorioretinal scarring.

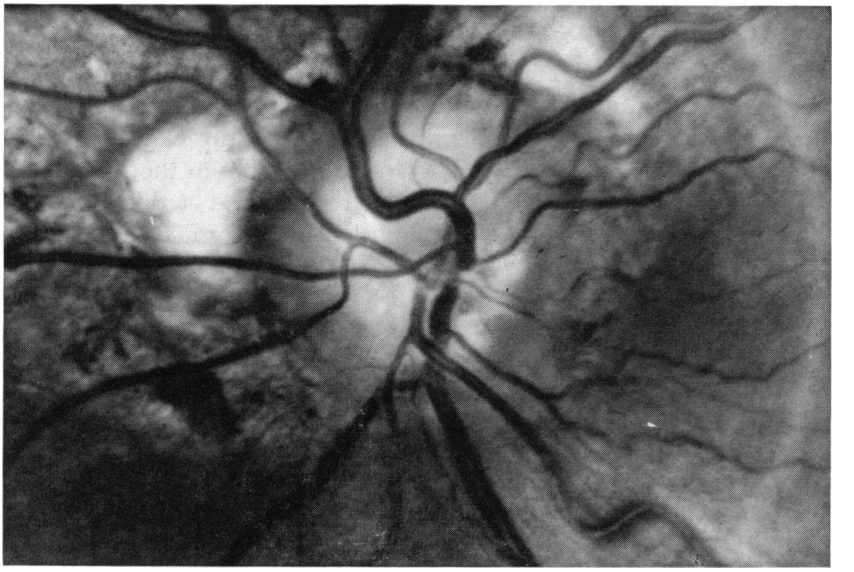

FIGURE 13B
(Case VII) Left eye: extensive peripapillary chorioretinal atrophy.

Comment

This is another example of subretinal hemorrhage associated with optic-nerve drüsen. The drüsen could be seen ophthalmoscopically, but their appearance was confirmed by a typical fluorescein angiogram, which also served to differentiate this condition from papilledema.

Six months later the hemorrhage had resolved and was replaced by peripapillary atrophy and pigment clumping similar to that seen in the left eye initially.

COMMENT

After evaluating the present series of seven cases and those scattered throughout the literature, it would appear that hemorrhage on and around the disk secondary to giant drüsen falls into three rather separate and distinct types: first, small hemorrhages in the nerve fibers of the disk and localized to the nerve head; second, hemorrhage on the nerve head extending into the vitreous; and third, deep peripapillary hemorrhage extending from the nerve head under the surrounding retina.

The first type, in which the hemorrhage is localized to the nerve

fibers on the disk, is probably much more common than would appear from the literature. Subjectively there is no visual loss, the small hemorrhages being chiefly noted in those patients undergoing a routine examination or being thought to have papilledema. These hemorrhages fade very rapidly and their finding ordinarily would be a coincidence. The importance of this type is probably in the differential diagnosis from papilledema. It should be noted as a differentiating point that, in papilledema, the hemorrhages are usually around the disk margin, whereas those secondary to drüsen are in the substance of the nerve head itself.

Hemorrhage extending into the vitreous is apparently the most common type noted in the literature and the first type to be described.[11] It is interesting to note that both Reese's patient[12] and one of our patients (Case II) had been under previous observation for drüsen and the hemorrhage was a late complication. Most of these patients will seek ophthalmological assistance because of the visual blurring resulting from the posterior vitreous opacity. As in our patient (Case II), this hemorrhage is probably absorbed, in the majority of patients, without visual loss, although there was possibly some permanent loss noted in one of our patients (Case IV). This specimen shows organization of the residual hemorrhage on the nerve head (Figure 5). The ophthalmoscopic appearance of this condition is very similar to that seen in subarachnoid hemorrhage. It is probably worth noting that the case reported by Gaynes and Towle[10] was associated with an attack of migraine, and in one of our patients (Case II), the hemorrhage appeared after strenuous exercise.

One of the most interesting aspects in the present series is the incidence of deep peripapillary subretinal hemorrhage secondary to drüsen in the disk. The only examples in the literature in which it was stated that there was "a minor peripapillary hemorrhage" are those of Gallais.[13] Apparently this condition has never been described as an entity in the literature.

Five of our patients (Cases I, III, V, VI, and VII) are of this type, and are therefore of particular interest. Although the ophthalmoscopic appearance varied in these five patients, all of them showed evidence of recent hemorrhage around the disk extending under the retina. All of these patients had subjective visual disturbances caused by the hemorrhage.

In two patients the hemorrhage was massive enough to cause a localized hematoma simulating a melanoma. In one patient (Case III) the eye was enucleated because of this error in diagnosis. This was

done some ten years ago, and with fluorescein angiography it is probable that the proper diagnosis would have been made. In one other patient (Case v), the possibility of a malignant melanoma was also entertained. With the appearance of the disks, not only on the side of the lesion but in the other eye, and with the characteristic field defect, the correct diagnosis was made without fluorescein angiography.

In three patients (Cases i, vi, and vii) the subretinal hemorrhage was diffuse and did not cause a localized hematoma. In two of these patients (Cases vi and vii), there was visual disturbance caused by macular distortion from the subretinal lesion. One of the patients (Case i) had no such macular disturbance; but coincidental with the hemorrhage there was a progressive field loss, to be described later.

In two patients (Cases v and vii), the vision returned to normal over a period of some months. However, in each example there was a permanent peripapillary pigment atrophy. In both of these patients there was a similar but less marked peripapillary atrophy on the opposite side, suggesting a previous similar lesion on that side without visual disturbance. It is possible that some cases of so-called peripapillary postinflammatory atrophy could originate in this type of lesion.

In all three types, it is believed that the hemorrhage originates from a direct effect on a blood vessel by drüsen. Drüsen are histologically hard, sharp concretions and with increasing size could erode a blood vessel. Also by direct compression on a vein, secondary venous rupture could occur.

The different types are apparently derived from variations in the vessel of origin, the extent of the hemorrhage, and its route of spread. In the type that is limited to the disk fibers, the hemorrhage is probably local and remains in the fibers of the nerve head. It is comparable to the fiber layer, flame type of hemorrhage in the retina. If the hemorrhage becomes more extensive, it can break through the hyaloid membrane into the vitreous, in a manner similar to the way extensive retinal hemorrhages spread internally. The deep peripapillary hemorrhage may arise from deeper vessels in the disk. The spread is from the substance of the disk under the retina between the rods and cones and the pigment epithelium. The route of spread is histologically demonstrated nicely in one patient (Case iii, Figure 4C). It should be noted that this lesion is different from subretinal disciform degeneration, where the hemorrhage is primarily under the pigment epithelium.

As noted, field defects are very commonly found in drüsen of the optic nerve head and, in many cases, are characteristic. Rucker[4] has

described three basic types: (1) concentric constriction, (2) enlargement of the blind spot, and (3) nerve–fiber-bundle defects. Harrington[15,16] has also suggested that all of these defects can be produced by drüsen. The high incidence of field defects in drüsen has been recently emphasized by Lorentzen,[1] who found an incidence of pathologic change in 79 of 91 examples (87 per cent).

Concentric contraction is apparently very infrequent, usually slight, and may not be related to the drüsen. Enlargement of the blind spot is commonly seen (60 per cent of Lorentzen's patients), and is usually marked with absolute borders. Nerve-fiber defects occur frequently, and some forms can be considered to be characteristic. Theoretically, three types may be present: (1) arcuate defects may present as a peripheral Roenne step, (2) nasal cuneate or wedge-shaped defects, and (3) centrocecal scotomata.

Arcuate defects are, of course, commonly seen in other conditions, being characteristic of glaucoma. They may be seen with drüsen in a very similar form. Cuneate defects are seen only rarely in other diseases such as juxtapapillary choroiditis, but are so commonly seen in drüsen that their presence should suggest this diagnosis. Even though the temporal half of the disk is commonly involved in drüsen, central defects are rarely noted. If this type is found, the presence of secondary hemorrhage should be suspected.

Four of our patients (Cases I, V, VI, and VII) had careful and repeated visual-field studies. All of these had hemorrhages of the deep peripapillary type. All four of these patients had nerve-fiber-bundle defects thought to be characteristic of drüsen.

In one patient (Case I), who had only one eye, there was marked progression of a pre-existing arcuate scotoma following a deep peripapillary hemorrhage. This progressed to a very small residual central field with marked reduction of central vision. This is one of the few examples in the literature in which there was an extreme visual loss from drüsen of the disk.

The remaining three patients (Cases V, VI, and VII) all had characteristic bilateral field defects even though the hemorrhage was unilateral. These bilateral defects were usually arcuate or cuneate with a defect secondary to the hemorrhage. The hemorrhage itself causes a rather typical enlargement of the blind spot with relative borders that would be expected from the pattern of hemorrhage in each of these cases.

As noted, the small splinter hemorrhages in the substance of the disk apparently have very little clinical significance. Theoretically,

there is one possible importance of this finding. Drance[17] has reported similar, if not identical, hemorrhages in patients with glaucoma. He has speculated about a possible causal relationship between these hemorrhages and the field loss occurring in glaucoma. If his theory is true in glaucoma, it could also be true with the similar hemorrhages found in drüsen.

As there is no treatment of the condition, the important clinical aspect is prompt recognition to avoid further unnecessary diagnostic procedures. The difficulties of differential diagnosis from papilledema have been described repeatedly.[18,19,20] Two patients (Cases II and VII) each previously had a mistaken diagnosis of papilledema. However, in our seven patients the presence of drüsen of the optic nerve head was easily confirmed by careful ophthalmoscopic examination. All of our patients who had visual fields had defects characteristic of drüsen but, in addition, the majority of them had central field defects secondary to the hemorrhage. Two patients (Cases I and II) show the tendency of drüsen to become more obvious clinically over a period of time. It is worth noting that drüsen of the disk with hemorrhage are often seen in young adults.

The use of fluorescein angiography in the differential diagnosis has recently been summarized by Sanders and fftyche of London.[21] In true papilledema, diffuse fluorescence of the disk and surrounding retina occurs with spread along the retinal vessels. In drüsen of the disk, the fluorescence is of irregular density, the margins discrete and nodular with no leakage along the vessels. Although only the last of our patients (Case VII) had fluorescein angiography, the findings were diagnostic. In addition, the peripapillary hemorrhage was obvious by retrofluorescence (Figure 12).

We wish to emphasize that in any patient, particularly a young one, in whom hemorrhage appears on or around the disk, the diagnosis of drüsen of the optic disk should be suspected. The disks, not only on the affected side, but on the opposite side should be carefully scrutinized for the presence of refractile bodies. Careful visual-field studies should be obtained. The use of fluorescein angiography is most desirable and will probably confirm the diagnosis.

SUMMARY

Drüsen of the optic nerve head are associated with three types of hemorrhage complications. One is the splinter hemorrhages in the substance of the disk. These apparently are asymptomatic, but

theoretically they may play a role in the field loss associated with drüsen. The main problem they present is in the differentiation of drüsen from papilledema. The location of the hemorrhages in the disk and the characteristic fluorescein angiography should serve to clarify the diagnosis.

The second type is the disk hemorrhage that is severe enough to rupture into the vitreous body. This results in blurred vision but usually resolves without complication. It is important to realize that drüsen of the optic nerve can be a cause of vitreous hemorrhage.

Third is the deep peripapillary hemorrhage lying beneath the retina, with associated blurred vision. It apparently resolves, leaving a peripapillary pigment atrophy similar to that occurring after inflammatory juxtapapillary processes. This type may also form a localized hematoma simulating melanoma. A central field defect is usually present in both forms. This report is apparently the first description of such a lesion.

ACKNOWLEDGMENTS

We wish to thank Dr Frank Winter for permission to include Case III, and Dr Allan Kolker for contributing Case V.

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DISCUSSION

Dr Thomas R. Hedges. The authors have added to nine patients in the world's literature seven from recent, personal, well-documented, experience wherein there were drusen associated with hemorrhage (1) superficial, on or next to the papilla, (2) into the vitreous, and (3) subretinal associated with striae of the retina or chorioretinal atrophy adjacent to the disk. These three groups bear careful scrutiny because their significance rests on whether or not the hemorrhages are more than coincidental findings.

Before receiving a copy of this paper, I reviewed the literature and the experience of many of our colleagues, with skepticism of a valid association between drusen and hemorrhage, considering migraine, other causes of bleeding or other vasomotor phenomena. I would agree the literature complements this unique series of patients. The paper directs our attention to three groups, about which I would like to comment.

(1) Superficial hemorrhage may or may not be directly due to or caused by an expansion of drusen. Since drusen are often difficult to detect when in or just anterior to the lamina, they pose a clinical problem in their differentiation from papilledema, and splinter hemorrhage only makes differentiation from choked disk due to increased intracranial pressure more difficult. When drusen are present with or without superficial splinter hemorrhage, the edict holds that the patient's other clinical symptoms and signs should dictate whether or not further studies are in order.

(2) Vitreous hemorrhage without associated cause brings us to the heart of the issue, namely whether the hemorrhage is truly caused by the drusen eroding vessels in the prelaminar area of the optic nerve. Neither I, nor anyone else, can prove if this is so; but the fact remains that both the authors and other esteemed colleagues feel they have documented such a possibility, particularly with giant drusen, which may

represent a hamartoma pathologically. In addition, drusen have, on rare occasion, been seen to develop and enlarge even leading to progressive field loss. Therefore, a pathologic sequence leading to hemorrhage is not unlikely.

(3) Subretinal hemorrhage with drusen really intrigues me, because there we have neither a superficial phenomenon, which could be coincidence, or a catastrophic one with vitreous hemorrhage.

This discusser's contribution to this group consists of one patient, discovered on routine examination, who had parallel retinal folds and drusen. Whether previous hemorrhage of consequence existed is speculative, although the same phenomenon occurring in both eyes leads to skepticism as to a relationship.

The second patient with drusen and minor subretinal peripapillary hemorrhage was contributed by Dr Weiner of Wilmington, Delaware, who kindly lent me these photographs. Here it would be doubtful if any defect will remain because the hemorrhage is so small.

I feel the authors have made a real contribution. When we see suspiciously elevated or full optic papilla with or without drusen which transilluminate by direct ophthalmoscopy or by fluorescein, we should give more than token recognition to a hemorrhage in or next to the nerve. This in the light of the dictum that drusen with or without hemorrhage should preclude further neurologic studies.

Vitreous and subretinal hemorrhage, though very rare, should be accepted clinically as associated anomalies wherein parallel retinal folds, other edema residues of oedema or peripapillary atrophic changes may occur.

In conclusion, I should like to present my own classification of drusen at the optic nerve:

(1) Idiopathic drusen not attendant by any visible pathology of the optic nerve other than the drusen themselves which may be either (a) nonfamilial (common), or familial (rare); (b) nonprogressive (common), progressive (rare), and regressive (?).

(2) Associated with other pathology, such as optic atrophy, vascular anomalies, retinal disease.

(3) Tuberous sclerosis.

I believe the author's presentation of drusen should be confined to group 1, which are idiopathic and not attendant by other pathology, because I believe the other groups only complicate the issue when we are talking about associated hemorrhages with drusen. One might add that congenital anomalies of the optic nerve, of which I have several associated with parietal retinal folds, should not be included in this discussion.

DR BEDELL. Spots, as small parts differing in color, finish, or material from the main part of the fundus may be sufficiently distinctive in form, distribution and life cycle to materially assist in the diagnosis of the ophthalmic lesion.

Blood, when enclosed in a dilated portion of a small blood vessel is a

microaneurysm. A typical one is a small artery. It grows slowly, rarely disappears spontaneously and is stationary. This was well seen in a 63-year-old woman with known diabetes for two years. An aneurysm was photographed over a period of many months and proved that there had been no alteration in size, color or location.

Similar aneurysms are found in congenital multiple gross aneurysms which look like a bunch of grapes so poorly nourished that many small ones are present.

In a large, irregular quadrate, macular mass of Coats' exudative choroiditis there were several isolated microaneurysms on small branches and on the sides of a large vessel.

In hemorrhage the color varies greatly from pale red to old, dark, homogeneous clots. A small retinal hemorrhage rarely has the smooth surface of an aneurysm and if followed long enough always shows either an increase or decrease in size with altered outline.

In contrast to blood or aneurysms there are pale sometimes translucent or yellowish spots beneath, or about, the retinal vessels, they may be scattered about the posterior pole, form a complete layer over the fundus, be widely separated or closely packed together. They are commonly called drusen, colloid bodies in Bruch's membrane, and are easily diagnosed by their location and color. Those in the elderly are almost white, several large vessel diameters in size and irregularly pigmented, the characteristic of Tay's Choroiditis.

It may be that large spots of greater thickness, more irregular distribution and seemingly deeper in the fundus, alluded to as choroidopathy, are pathologically similar to the ordinary drusen but visual disturbance is greater.

White spots in the fundus may be few or so numerous as to practically cover the entire visible background and be associated with pigmentations usually of the bone corpuscle shape and nightblindness. The age of onset is variable.

A woman who died when she was 81 years old had been observed for 40 years. The dominant features of her fundi were the great number of separate, white spots beneath the retinal vessel and an occasional branched pigmentation. She had always been nightblind. A classical picture of retinitis punctata albescens with retinitis pigmentosa. As time passed, the discrete, white spots disappeared in areas of choroidal atrophy and pigmentations in the equatorial and peripapillary regions.

In others with nightblindness, the white spots are fewer, new ones develop more slowly and pigmentations are present, retinitis punctata albescens.

The female type of choroideremia has dark, almost black, spherical spots in the equatorial zone with nightblindness.

The soft edged, pale, rounded areas found in conjunction with malignant melanoma of the choroid suggest metastases by their increase in size as well as their distribution about the growth.

Flat, reddish hemorrhages in malignant melanoma may be the cause of a sudden reduction in vision.

The dispersion of pigment in senile macular exudative stages ranges from minute specks to those several times the width of the major retinal vessels. They are flat, comparatively thin and as time passes become measurably smaller and lighter colored.

In traumatic retinochoroiditis the areas of pigmentation are usually dark brown or black, small or large plaques, which increase in size and number for a time after the injury and then become stationary. Rupture of the choroid is a common cause.

Prolific sources of yellowish white and whitish yellow spots in the fundus are diabetes, nephritis, and hypertension.

The yellow white spots of diabetic retinopathy with microaneurysms have been known for years. They may develop from small specks and become large masses with hemorrhages both early and late. The retinopathy usually appears in those who have had diabetes for years and have been under "control" by diet, insulin, or new drugs.

After sudden occlusion of the central retinal vein delicate, minute, yellow dots may be among the late signs.

In nephritis the spots may be soft, gray white edemas, ischemic areas often in stellate groups about the macula. Those in hypertension are always accompanied by visible vessel alterations.

Senile macular degeneration may start with small hemorrhages. During the course of the disease pigmentations are common.

Cholesterin crystals are easily recognized by their iridescence.

The congenital pigmentations are flat, irregular in outline and size with the larger ones nearer the equator, they usually remain unchanged.

DR PHILIP M. LEWIS. I personally have not seen hemorrhages in connection with drusen.

I would like to ask the essayists, as they reported seven cases, how many drusen they actually see, and percentage-wise how many do have hemorrhages? I also wonder if sometimes these hemorrhages are not due to other related causes and not to the drusen.

Third, I would like to comment that I have never seen drusen in the Negro race. I would like to know what the experience in that regard has been.

As drusen may exist in patients who have an optic neuritis or an intracranial lesion, I think it behooves us to think of the possibility of this, and not simply always conclude that the drusen are solely responsible. I would like to know how they know which cases to investigate, provided there is depression of central vision and progressive disease.

Such a case I report briefly.

A young man was seen 12 years ago at age 31 with bilateral drusen. He came in because of depressed vision of his right eye. Vision was down to 20/50 with considerable field changes. Feeling that he might have

something else, I referred him to a neurosurgeon who did ventriculograms and decided that the reduction was probably due just to the drusen. I saw him a couple of years later. Then he came in this year. He is very much the same, except that central vision of the right eye has fallen off considerably.

[Slides] These slides show his two disks and the visual fields in 1958 and then in 1970.

How can you tell which cases should be referred to a neurosurgeon and which should not? Should you assume the entire responsibility just because you think it is entirely drusen, or should you get a little extra help? I have followed this case for twelve years. If any of you are still around in another ten years I will make another report on it.

DR KEARNS. In the absence of Dr David Cogan, and without his permission, I would like to mention a patient whom he saw last November. I feel I am justified in reporting this case, in that I was in Boston and Dr Cogan kindly showed me this patient.

The patient was perfectly healthy, about 30 or 35 years of age, as I recall. The only finding was a large hemorrhage on the right optic disk. I do not recall the exact appearance, but it was a significant-looking hemorrhage. Dr Cogan thought that it was caused by drusen and not by choroiditis or some similar cause.

I checked with my colleague, Dr Robert Hollenhorst, and in our institution in recent years we have not seen a hemorrhage associated with drusen. Dr Rucker does not think we saw them in previous years, either. Such hemorrhages must be rare, or all patients having them go to St. Louis.

There is one other point in the paper that I think is worth emphasizing, and that is the loss of central vision. We used to think that patients with drusen might have severe peripheral loss of vision, but if they had loss of central vision it was not due to hyaline bodies. In the last few years Dr Hollenhorst and I have seen two patients who, we think, definitely had loss of central vision caused by hyaline bodies. I am interested that at least one of the patients described by Dr Gay had loss of central vision.

DR SWAN. Occasionally the hemorrhage can be so large that it masks the drusen. If the patient is one-eyed the differential diagnosis becomes difficult, as illustrated by the following case:

A 22-year-old logger had lost one eye in an accident years before. In 1968 he had a sudden blurring of vision and was referred to the University of Oregon Medical School with a diagnosis of occlusion of the central vein. We were asked to consider the possibility of anticoagulant therapy. The referring ophthalmologist also considered papilledema and optic neuritis.

Upon admission examination, visual acuity was 20/30. Quantitative perimetry revealed an irregular depression of the visual field and enlargement of the blindspot. He had a normal physical examination, including neurologic consultation, radiologic studies and angiography.

[Slide] This was the initial appearance of the fundus. Notice the characteristic subretinal hemorrhage to which the essayist called our attention. There also was bleeding into the vitreous producing some haze. Fluorescein angiography made it unlikely that there was papilledema or pappilitis. Pulsation of the central vein was visible, which excluded venous occlusion. Diagnosis of hemorrhage associated with drusen was made. No treatment was undertaken.

[Slide] This was taken six months later, showing the drusen quite clearly. A year and a half later there has been no recurrence of the bleeding.

In this case the establishment of the diagnosis saved the patient from extensive neurosurgical studies and the dangers of anticoagulant treatment.

DR MAUMENEE. Prior discussants questioned whether there was a real relationship between hemorrhage and drusen of the disk. I want to report that I have seen two children under age 15 with hemorrhagic detachment of the macula, with drusen of the optic nerve.

DR LARRY L. CALKINS. Just one additional comment in relation to Dr Maumenee's report. I have followed a boy, now age 16, over the last five years with exactly the same sequence of events. He was seen at age 2½ by Dr A. N. Lemoine, Jr. with an esotropia, and the comment at that time was that there were abnormalities of the nerve head probably congenital in nature.

The nature of the optic nerve lesion was not obvious for several years, but now both optic nerves have drusen. He has had repeated peripapillary subretinal hemorrhages finally involving the macula in one eye, with reduction in vision to finger counting. The other eye has a large cecal scotoma. He is a crack basketball player and top guard on his team, with no stereopsis.

The second patient was a 36-year-old woman referred to me in 1956, with bilateral drusen, rather large ones, and gliosis as described by Dr Gay, which I presumed might have been previous hemorrhage. She subsequently has shown repeated hemorrhage and extension of her cecal blind spots to involve first the macula of her right eye, with marked reduction in vision, and within the last year the macula of the second eye. She now has finger counting in both eyes.

DR FRANK B. WALSH. Both our President and I have mixed up brain tumors and hyaline bodies in the optic nerve.

Some years ago I made this mistake, and the patient was found to have not one but seven intracranial meningiomas. Dr Okun reported on my error and I will accept it as such. Recently I have seen a patient with obvious hyaline bodies and equally obvious increased intracranial pressure. So, I have had an opportunity to see a patient who had hyaline bodies, increase in intracranial pressure, and in whom the tumor was successfully removed. The disks became flat.

I had difficulty in being sure that I could recognize the hyaline bodies,

which is important. I did recognize them. I have seen hemorrhages with hyaline bodies, but sufficient has been said about that.

DR SMITH. [Slide] Here is a young boy with typical optic nerve drusen. This is his left eye.

[Slide] He recently had an onset of blurred vision in the right eye, and showed both these superficial striae and the nasal margin hemorrhages. I thought this was a very similar case to the one Dr Gay presented.

[Slide] This is the same eye with fluorescein.

[Slide] This is Dr Gass's second case. This was a 21-year-old patient who also presented with evidence of old peripapillary inflammatory or pigmentary change.

[Slide] This shows a small hemorrhage. This patient had a skin test. We must recall the frequency of peripapillary inflammatory change in histoplasmosis. We must differentiate the primary cases, as Dr Hedges pointed out, that have optic nerve drusen alone, from those that have secondary changes due to coexisting eye disease with optic nerve drusen.

[Slide] This is a fluorescein picture of the last case.

[Slide] I had a case at Duke, in 1961, of a young lady who developed preeclampsia late in her pregnancy. She was found to have hemorrhage on the edge of the disk and was thought to have papilledema. She was sent to Duke for consultation to see whether or not the pregnancy should be interrupted. She had a choked-appearing disk with a hemorrhage on it. It was thought then to be papilledema. Further inspection showed this was a hyaline body with splinter hemorrhage on the edge. Three months later the hemorrhage had cleared.

[Slide] This is a patient we have followed, to show you the extent of the visual loss and field defect that can occur. She had 20/25 acuity in one eye and 14/200 in the other eye, with extensive nerve fiber bundle field defects. [Slide] This shows her disks in 1963. [Slide] Again in 1967. [Slide] On a 4-year follow-up in this case there was no significant change in the disks or drusen.

I have pictures of drusen patients who have uniocular field loss, with only one quadrant remaining in the other eye, yet with good vision. I have another case of 20/15 in one eye and only one quadrant in the other eye, with poor central vision, with 5-year follow-up.

Another neurologically important point was made in Dr Gay's last two cases. A woman was referred by Dr Isbey from Asheville, North Carolina, with drusen induced field defects simulating a left optic tract hemianopia. You can see the same suggestion in Dr Gay's cases 6 and 7, for the lower nasal field defect in the left eye corresponded with an incongruous lower temporal nerve fiber bundle defect in the other eye.

With these blurred disks one must proceed carefully. Dr Isbey's patient had carotid arteriograms and an air study, and these were negative. She had a field defect mimicking an optic tract lesion; however, one year later there was no change. It is obvious that we now must be much more care-

ful with these patients than heretofore. Dr Hoyt says that the best way to tell your neurological colleagues about optic nerve drusen is to consider such a person as a patient with bilateral mature cataracts. Think of the patient as though you could not see the fundus. If you have an adequate indication for contrast studies, other than from the fundus, then you can proceed.

DR T. E. SANDERS. I want to thank Dr Hedges for classifying this condition. I think we all know now what we are talking about.

Dr Bedell, I expected someone to take exception to the causal relationship of these two conditions. There are several reasons why I think they are causally related. In the first type the hemorrhage is in the substance of the disk. In another type it spreads into the vitreous; in the third type it spreads under the retina, but all originate in the disk. I know of no other condition that causes intrapapillary hemorrhage.

Secondly, although we have only two cases (the sections Dr Gay showed you), we have very good histologic evidence that this condition exists. Being a pathologist, I like this.

Thirdly, these are all young people. Five of them were under age 32, in perfect health, with nothing else wrong with them as far as we could discover.

Fourth, they all had drusen of the disk and hemorrhage. As these are two relatively rare conditions, when you see them both together you have to think there may be some causal relationship.

On the other hand, Dr Bedell did not suggest any other cause for the hemorrhage.

To answer Dr Lewis's question, I don't know the incidence. We have just been interested in the ones with hemorrhage because these are the ones that have struck us. They were all white; we had no negroes.

Dr Smith answered his other question very nicely: What do you do with these patients when you see them with a splinter hemorrhage, with a presumptive diagnosis of papilledema? First, fluorescein angiography is going to be very helpful. Then, unless you have other neurological signs of intracranial disease, you watch them. Maybe you watch them with a neurosurgeon, but you sort of hold his hand.

I had hoped that our members would present one or two cases of this condition to substantiate its existence. I had not hoped we would have had so many presented. I want to thank all of you for your interest in bringing these cases here.

I would like to close with an historical note. The first case of drusen of the optic disk reported in the United States was presented to this Society in 1892 by Dr deSchweintz. The first hemorrhage associated with drusen was described in the literature in 1895 by the father of one of our members, Dr Harold Gifford. It was not until 1940 that the second case was described by Dr Algernon Reese.